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Two Unique Cases of Congenital Occlusion of the Anterior Nares.

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TWO UNIQUE CASES OF CONGENITAL OCCLUSION OF THE ANTERIOR NARES.*

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Complete congenital stenosis of the anterior nasal orifices of the nature I am about to describe must evidently be viewed as an extremely rare condition, inasmuch as, after a careful and conscientious search, I have been unable to find a single case reported in the medical literature of the subject, and the two reported in this paper were the only ones discovered among more than ten thousand patients recorded at my university and Bellevue Hospital clinics, and in private practice. This observation and the fact that I am able to present a case of imperforate nostrils successfully treated by means of my nasal drills and an electric motor, re-enforced by an invitation from our esteemed president to read a paper before this association, are the principal motives which have induced me to infringe upon your valuable time.

The first case to which I shall invite your attention is that of a young man, eighteen years of age, who, though

* Read before the American Laryngological Association at its ninth annual congress.

unprepossessing in appearance, showed a fair state of physical development.

He first consulted me in September, 1885, in search of relief from a life-long closure of both nostrils. The gentleman to whom I was indebted for this most interesting case stated that his attention was particularly attracted to the patient's condition by reason of an unfavorable prognosis pronounced in his hearing by a well-known surgeon, which was substantially that the condition of the young man's nose was irremediable, and that he had better submit to the annoying, though not serious, discomfort rather than undergo an operation the successful issue of which was involved in much doubt, a view which appeared quite plausible on making a superficial examination.

On ordinary inspection, the dark outlines of the anterior nasal orifices were seen to be replaced by two cup-shaped depressions about four millimetres in depth. This barrier consisted of a dense, white, glistening membranous wall, fringing the inner face of the alæ nasi, and fusing with the side of the septum lying opposite. The center of the cup-shaped depression in the right nostril was perfectly *smooth*, not being marked by so much as a wrinkle. A close inspection of the left anterior nasal orifice (Fig. 1) revealed the presence of a diminutive opening, *a*, at its upper portion, capable of accommodating with difficulty the point of a fine probe.

The external nose was large and well developed, excepting an insignificant transverse crease on the right side. An interesting feature was the striking enlargement of both the upper and lower lips. This peculiarity would appear to afford corroborative evidence of the accuracy of Ziem's view that labial hypertrophy might be present as a result of permanent nasal stenosis. The practice of posterior rhinoscopy was rendered quite difficult, but I finally succeeded in obtaining a satisfactory detailed sketch of both posterior nares.

The vault of the pharynx appeared perfectly free. Each post-nasal opening was distinctly visible; the right one, however, was seen to be imperforate at a point beginning about three eighths of an inch from the free edge of the vomer. The

cause of this occlusion was, furthermore, distinctly visible as an extreme deflection of the vomer to the right, even to the point of contact with the opposite nasal wall.

The amplitude of the left choana was naturally increased at the expense of the right post-nasal opening, and it was possible to make out the left inferior turbinated body, although it appeared smaller than usual.

This brief outline will convey a general idea of the anterior and posterior rhinoscopic appearances.

In addition to the patient's personal record of a life-long discomfort from complete stoppage of the nostrils, I was fortunately able to obtain a detailed biography of the individual from the time of his birth.

This information was drawn from the young man's mother, and it might be added to her credit that she proved to be an intelligent and painstaking observer.

I have extracted the following brief notes from the *body* of her narrative, as constituting important evidence of the congenital character of the deformity: About six weeks after the birth of the child her attention was attracted to the infant's nostrils by a slight discharge. Shortly after making this observation she also observed that the child breathed with great difficulty through the nose. She directed the attention of her physician to the nasal difficulty, and, in accordance with his instructions, essayed to syringe the nostril, but her effort in this direction proved futile, for the reason that the fluid immediately rebounded on being projected into the nostrils. Snuff was then prescribed, but it was found impossible to sneeze away the obstruction. During the first year of the child's life she remembers having seen the pin-hole perforation already referred to as occupying the left nostril of the young man. From the moment she first noticed the obstruction in breathing, during the infancy of her boy up to the time of her application to me for relief, her son had never drawn a breath through the nostrils. The misery produced by this condition has proved the burden of her life. Ever and anon has she been aroused from her slumbers by the suffocative sounds made by her son struggling in his sleep for more air.

During these paroxysms the boy's tongue appeared to fall backward into the throat and so block up the only available avenue for the entrance of air into the lungs. It required violent shaking to arouse him from the stupor which accompanied these attacks, which, like similar manifestations observed in children suffering from the obstructive effects of enlarged tonsils, might be attributed to the carbonization of the blood resulting from deficient aeration of the inspired air.

As bearing upon the possibly scrofulous character of this anomalous condition of the nostril, the following notes, given to me by the mother concerning her son's state of health and that of her several children, may prove of interest. Mrs. F. recollects having suffered during her childhood with a nasal difficulty. The present condition of her nostril clearly demonstrates the pre-existence of either scrofulous or syphilitic disease. The external nose is in a state of collapse, a deep furrow running transversely across the dorsum at the level of the nasal bone. The tip of the nose is tilted upward, and anterior rhinoscopic examination demonstrated the almost entire disappearance of the cartilage of the septum, along with a considerable portion of the vomer and plate of the ethmoid bone. Both nasal cavities are abnormally spacious. She has borne seven children in all. Her first child had been subject to attacks of nose-bleed. One day, while at play, a ball accidentally crushed his nostril, provoking a profuse and persistent hæmorrhage, which only ceased with the child's life. The attending physician attributed the fatal result to a ruptured blood-vessel. Another son was subject to severe headaches, which were often accompanied by nose-bleed. He is now fifteen years old and a victim to catarrh. Only three of her children apparently possessed good health.

Diagnostic Conclusions.—Basing my deductions upon the foregoing history and appearances, I would offer the following explanation to account for the origin of this remarkable nasal anomaly: The individual was probably, in the first place, born with a malformed nostril, the evidence of which exists in the form of the misplaced vomer, which

could not assume its faulty position either from injury or disease. The right nostril was probably occluded anteriorly by a membranous wall at birth; the left nostril, perfectly free posteriorly, communicated, by means of a small orifice in the anterior nares, with the outside world. Possibly this was cicatricial.

A scrofulous coryza, occurring at or shortly after birth, was the probable cause for the discovery of the catarrhal flux and subsequent stoppage of the nasal passage. Contraction of this small orifice of escape evidently rapidly occurred until it was reduced to a mere pin-hole perforation.

The surgical treatment adopted for the correction of the condition just described was commenced on April 26, 1886, the first operation being performed at the University Medical College, before the students. A perforating drill having a quadrilateral cutting face, the four knives being arranged at right angles to each other, was employed to make the preliminary puncture through the dense fibrous membrane. This drill was propelled by an original modification of the common surgical engine. Weber's drill chuck was employed. The right cup-shaped depression having been sprayed with a strong solution of cocaine, the perforating drill, revolving at full speed, was directed against the obstruction, through which it easily made its way, reaching the free space in the deeper portion of the nostril. The instrument being then withdrawn, my rasp drill was introduced within the preliminary opening, which it promptly and freely enlarged laterally. At the conclusion of the cutting, rhinometric measurements showed the transverse diameter of the newly made nasal orifice to be just four millimetres, and the vertical measurement six millimetres. The slight hæmorrhage which followed the practice of this procedure was readily controlled by means of a pledget of cotton and did not inconveniently impede the progress of the operation. A free rush of air through the new nasal orifice convinced us of the success of the operation. The edges of the incised obstructing mem-

brane appeared to be about two millimetres in thickness. On a subsequent occasion the right nostril was laid open by a repetition of the above procedure, with slight modifications. My patient, who had been formerly dismissed as cured, called upon me again in April, 1887. An examination of his nostril showed that, whereas the left anterior naris had remained patent, the other nostril had contracted to the size of a useless slit. He had realized so much comfort, however, from the use of his free nostril that he felt impelled to seek relief through another operation for the restoration of the right nostril. In reopening this nostril, I cut more freely with my drills than previously, removing the dense fibrous tissue up to the point of its attachment to the osseous walls of the nose. To insure perfect success, I took advantage of the opportunity and enlarged the already patent nostril to the extent of several millimetres.

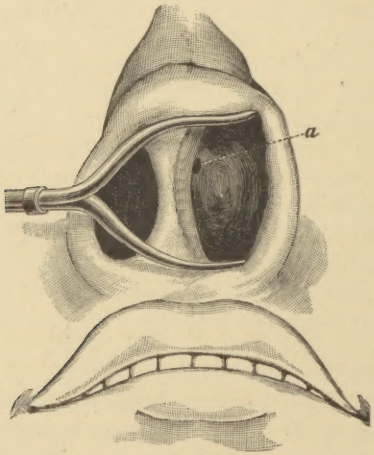

FIG. 1.

These sketches, taken from life, will enable you to appreciate the appearance of the nostrils before and after the performance of the several operations practiced for the relief of this unique and distressing malformation.

Electricity was utilized to obtain the motive force for propelling the drill during the second operation. The mo-

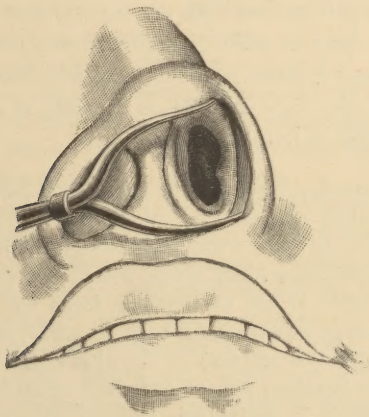

FIG. 2.

tor employed by me is known as the C and C motor. Different from the instruments of this kind usually found in the market, the C and C motor is supplied from a single quantity cell or from any ordinary galvano-cautery battery. The armature, which connects directly with the drill, revolves between fifteen hundred and two thousand times a minute. The motor and flexible shaft are manipulated while suspended in mid-air. The motions of the armatures are concealed by means of a metallic shell, which does away with the terrifying effects excited by the sight of the rapidly revolving axles.

By means of an adjustable connecting-rod I am enabled to attach the flexible cable to either side of the armature. It is obvious that this device permits me to use either right or left cutting drills. The force exerted by the motor is estimated as high as one eighth horse-power.

I also take this opportunity to present two other forms of my tubular drills, which I have termed, respectively, a spiral and interrupted spiral. They are better adapted for cutting through soft tissues than my tubular rasp drill.

The next case to which I shall direct your attention is that of a young woman, aged sixteen, who was brought to me by her step-mother in 1884 on account of an almost absolute stoppage of both nostrils.

As in the case just reported, the nose of this patient possessed neither respiratory nor olfactory value. The girl had been able at times to forcibly expel a little air through the nose. Despite the fact that the patient had never, within the memory of the mother, been able to use the nostrils, there was no deafness. The good hearing possessed by both of these patients would seem to disprove the accuracy of Toynbee's and Lucae's views concerning the injurious tension exercised upon the drum-heads during deglutition in individuals afflicted with nasal stenosis. The general health of the patient was poor. Her intellectuality, I believe, would be placed considerably below that possessed by young women of the same age. Whether this was due to the faulty conformation of her skull or was simply the natural intellectual dullness sometimes observed in individuals afflicted with long-standing nasal stenosis, or both, was an interesting though unimportant question.

Examination.—The first thing particularly noticeable about the young woman was the remarkable expression of her face. This peculiarity resulted from a variety of facial irregularities, conspicuous among which I might mention a flat, retreating forehead, an unusually broad interocular space, and the existence of two extraordinary prominences of the malar bones near each inner canthus. Add to this a drooping lower jaw, and you have a pen-picture of the unsightly physiognomy inherited by this unfortunate girl.

An examination of the anterior nares revealed the cause of the nasal stenosis in the shape of two pale-pink protuberances which appeared to completely block up each inferior meatus.

The columna of the septal structures was intact, and no portion appeared to be involved in the deformity. My first impression that the two obstructing bodies were extensive turbinated hypertrophies was quickly dispelled upon touching them with a probe. In reality, they were composed of bone, which I afterward discovered was exceedingly dense in consistence. The mucous membrane covering the structures was exceedingly thin. These bodies impinged directly against either side of the septum, and it was found impossible to introduce a probe through the narrow crevice found at the point of contact.

An examination of the naso-pharynx revealed the presence of nothing abnormal in character, and the posterior nares appeared to be spacious and well shaped. Basing my deductions upon the gross appearances just given, and the structural conditions revealed while operating, I felt justified in regarding the formation as a congenital abnormity of the principal bones of the face associated with marked malformation of portions of the anterior nasal framework, especially the inferior turbinated bones.

The bony structure of the turbinated bodies possessed more the character of an eburnation than the common osseous shell. Its density was so great that the points of the rongeur forceps frequently clashed together without cutting the bone, although I had succeeded with the same instrument in readily removing ordinary turbinated osseous tissue. These constituted the principal peculiarities of this unusual deformity.

Inasmuch as my surgical efforts did not yield the satisfactory results hoped for, I will not detain you with the details of the operation undertaken for the relief of the patient. It might be worth stating that while she was under the influence of chloroform a channel was cut through the patient's nostril as far as the naso-pharynx, my rongeur forceps being employed for the purpose. The immediate good result—restoration of nasal respiration—proved delusive, inasmuch as the contraction which followed the healing process soon robbed us of the respiratory space we had

acquired. Although my proposition to repeat the procedure was not received with favor, I am nevertheless confident that the employment of the electric drill in conjunction with intra-nasal dilators would have been followed by excellent results.

In concluding my remarks, I desire to notice the general significance of congenital occlusion of the nasal passages. This form of nasal stenosis is always referred to as an extremely rare condition. References on the subject are almost exclusively applied to abnormities situated in the choanæ; or, in other words, in the most obscure and inaccessible portions of the nasal cavities. The majority of these cases were reported as imperforate, the obstruction having the form of a bony plate, which spread like a web over one or both post-nasal orifices, and was directly continuous with the palate bone, of which it formed an integral part. Such a deformity, though demonstrated by a post-mortem examination to be possible, must nevertheless be viewed as almost unique in its occurrence. Though thoroughly convinced of the perfect sincerity of those who have from time to time reported cases of congenital occlusion, I feel, nevertheless, constrained to doubt the accuracy of their conscientious diagnoses.

Furthermore, I may add that I think any one is justified in taking exception to my own or any rhinoscopic diagnosis based upon an examination conducted by means of the finger in place of the mirror. Twenty recorded cases of congenital closure of the posterior nares in otherwise normal noses, many of which were double-sided, and not one of complete bilateral congenital occlusion of the anterior nares, is a disproportion that might well provoke surprise, or even incredulity. An inquiry concerning the unequal existence of these two conditions is most desirable. As one explanation, the strong probability of deflection of the vomer being mis-

taken for the plates of Luschka might be safely advanced. Congenital deflection of the vomer is a very common intranasal deformity. I have seen the misplaced vomer deflected to the point of contact with the lateral nasal wall. It requires no stretch of the imagination to picture an investigator, whose mind has been preoccupied with the idea of the existence of transverse congenital plates, thrusting his finger against an extreme deflection of the vomer and ratifying this preconceived impression. I consider a diagnosis of congenital transverse osseous plates or membranous occlusion of the choanæ, based upon digital exploration or the use of the probe, little better than mere guess-work. A careful and successful rhinoscopic examination should constitute the only criterion for the formation of such an important diagnosis.

Congenital deflection of the osseous septum has been shown to be an exceedingly common deformity, and therefore requires no especial mention in this connection. I have already considered its hereditary and pathological significance in a paper devoted to the ætiological relations between the malformed septum and pulmonary disease. The septum of heredity is almost invariably associated with an elevation of the roof of the mouth, which can often be traced back to a parental origin.

Finally, in order to indicate the scope of my unsuccessful search for the record of instances of congenital occlusion similar to those just described by me, I might mention the discovery of an interesting case of unilateral anterior stenosis in a cadaver dissected by Delstanche, which proved to be the nearest approach to my own. The left nostril was reported to be intact. The right nostril was imperforate, the alæ nasi being absent. Delstanche states that the condition was either congenital or acquired, and he favored the view of its being an inherited condition. It

is to be regretted that the most important testimony in this case, the life-history of the individual, could not be obtained.

Instances of acquired stenosis of the nasal passages are exceedingly common, and therefore possess no particular interest for us in this connection. As is well known, they may result from adhesions incident to injuries, ulcerative processes, and abrasions of the septum.

The cicatricial adherence of the septum to the turbinated tissues is a common condition, and often occurs as the result of the free and careless use of caustics, the galvano-cautery, and unskillful operative attempts to remedy the deflected septum. These forms of acquired atresia are merely mentioned to render the presentation of the subject more complete, and to show that they have been properly excluded from the category of congenital occlusions of the anterior nares.

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